Juvenile Rheumatoid Arthritis

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Juvenile Rheumatoid Arthritis

Overview

- Chronic synovitis in children
- First described in 1897 by George Frederick Still
- Several distinct subgroups
- Also called Juvenile Chronic Arthritis in Europe

Juvenile Rheumatoid Arthritis

Overview

- Pauciarticular
  - Girls > Boys
  - ANA positive vs. ANA negative
  - Chronic Uveitis 3 times as common in ANA positive
- Polyarticular
  - RF positive = Worse Prognosis
- Systemic Onset
  - ANA and RF are always negative
  - Chronic disease is unusual and worrisome

Juvenile Rheumatoid Arthritis

- Chronic autoimmune disease of unknown etiology
- Inflammatory process - systemic, joints, eyes
- Mediated by IL1, IL2, IL6, TNF, etc
- Antigens not yet identified
- Genetic component: general population risk 1 in 5-10,000, sibling risk 1 in 100

Pauciarticular JRA

- Most common type (40-50%)
- Subacute onset of pain, swelling, loss of function
- Girls 3 times more often than boys
- Peak age of onset 1 to 5 years (early onset)
- Knee most often, followed by ankle, wrist, elbow

- Late onset, after age 8, predominately males, is a subgroup perhaps related to spondyloarthropaty (HLA-B27 positive in 75%)

Pauciarticular JRA
- continued -

- Characterized by morning stiffness and gelling
- Exam shows joint swelling, tenderness, decrease in motion
- Diagnosis requires 6 weeks of persistent swelling

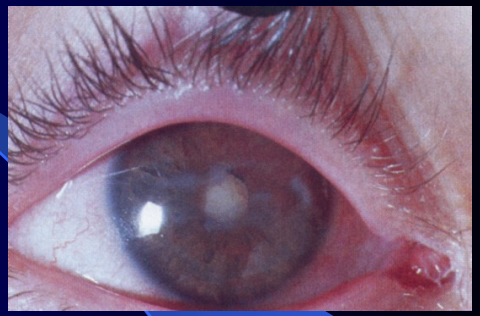
Pauciarticular JRA
Lab Studies

- Elevated erythrocyte sedimentation rate
- Normal CBC or mild anemia
- ANA positive in 30-60%
- RF is always negative
- LDH (for differentiating malignancy, see next slide)

Pauciarticular JRA
Differential Diagnosis

- Septic arthritis - characterized by severe pain, acute onset, and fever
- Malignancy - characterized by ill-defined but severe pain with an elevated LDH
- The rest are usually going to be transient or reactive arthritis, characterized by preceding illness, fluctuating course, and normal sed rate. These resolve in 6 weeks or less.

Pauciarticular JRA
Complications



Chronic uveitis (30%)

- Common but asymptomatic
- Three times as likely in ANA positive
- Slit-lamp exam on ANA positive every 3 months for four years
- Slit-lamp exam on ANA negative (10% risk) every 6 months for four years

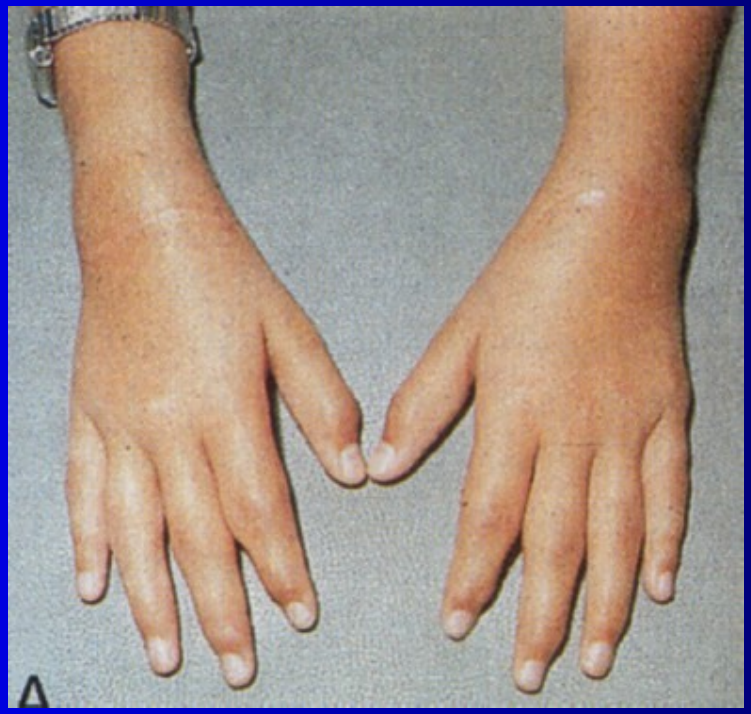
Pauciarticular JRA
Prognosis

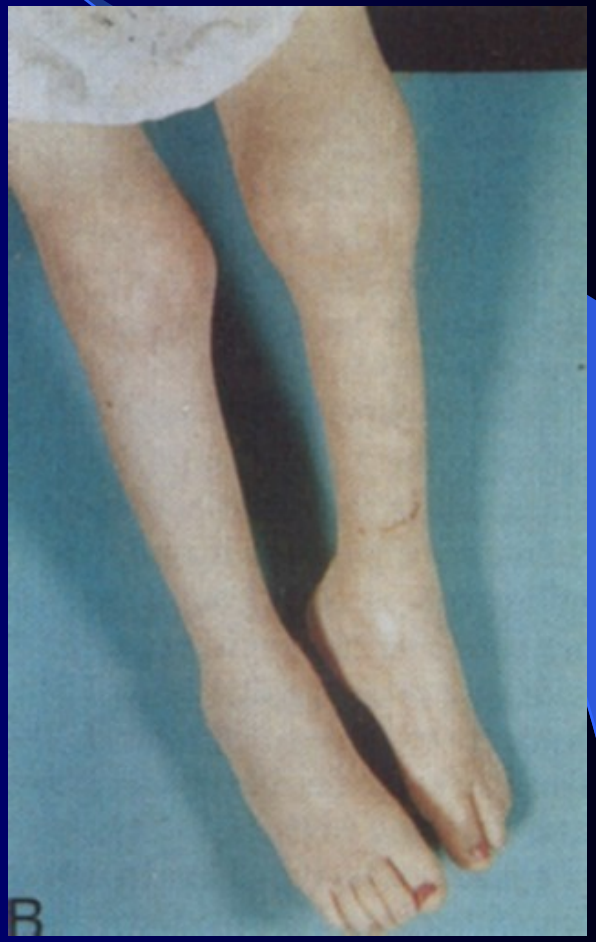
- Usually remits
- Seldom produces joint damage

Polyarticular JRA

- Second most-common type
- Symmetric involvement of small and large joints
- Subacute onset, does not migrate
- Loss of motion may occur rapidly
- Rheumatoid nodules are occasionally seen
- Subtypes: RF negative and RF positive

Polyarticular JRA





Polyarticular JRA
Lab Studies

- Elevated erythrocyte sedimentation rate
- Anemia is frequently significant
- RF may be positive in older children (typically over 8), may indicate more aggressive disease

Polyarticular JRA
Differential Diagnosis

- SLE - characterized by multi-organ disease with multiple autoantibodies
- Rheumatic Fever - Characterized by a heart murmur and a migratory pattern
- Parvovisus B19 - Characterized by a normal sed rate and a history of preceding exposure to Fifth Disease

Polyarticular JRA
Prognosis

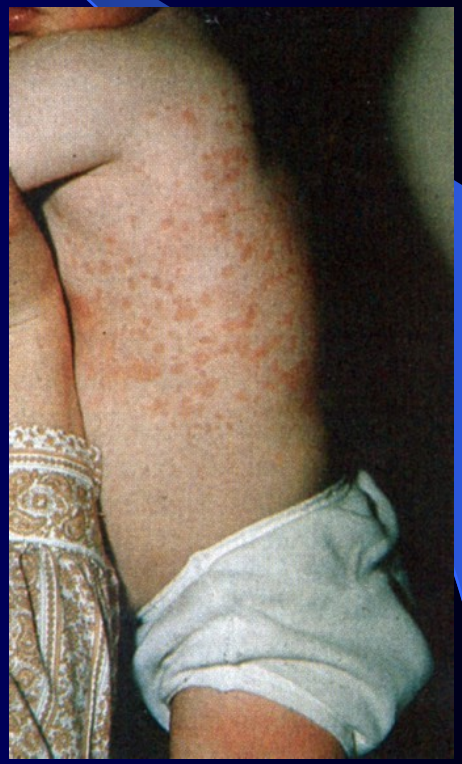
- Polyarticular with positive RF is worrisome
- RF positive can be rapidly progressive with joint destruction and permanent disability

Systemic Onset JRA (Still's Disease)

- Acute onset with daily fevers for weeks
- Diagnostic evanescent rash
- Variable joint involvement
- Markedly inflammatory lab studies
- Fever is characteristically daily or twice-daily, to 39° C or higher with intervening afebrile or subnormal temperature

Systemic Onset JRA
(Still's Disease)
- continued -

- Rash - small macules most commonly on trunk and proximal extremities, present during fever
- Myalgias and arthralgias are common
- Pericarditis and other inflammation of serosal-lined surfaces are common



Systemic Onset JRA (Still's Disease)

Lab Studies

Labs are highly inflammatory

- CBC shows leukocytosis with a left shift, thrombocytosis, and anemia
- Erythrocyte sedimentation rate is markedly elevated
- A normal ESR suggests another diagnosis
- ANA and RF are always negative

Systemic Onset JRA (Still's Disease)
Differential Diagnosis

- Malignancy - suggested by a high LDH
- Viral infection - suggested by a normal ESR
- Kawasaki Disease - criteria include desquamation of rash (hands, feet, groin), swollen lymph nodes +/- spleen, conjunctivitis, chelitis
- Other causes of FUO - may require extensive investigation

Systemic Onset JRA (Still's Disease)

Prognosis

- Usually monocyclic or polycyclic
- May be chronic - unusual and worrisome

Juvenile Rheumatoid Arthritis

Treatment

- NSAIDs are first-line drug of choice
- Methotrexate for recalcitrant disease
- Steroids - low dose for pauciarticular and polyarticular disease
- Steroids - high dose (1-2 mg/kg/d) in systemic disease

Juvenile Rheumatoid Arthritis

Pediatric Doses of NSAIDs

Drug	Dose	# doses/day	Comment	Cost
Ibuprofen	30-40 mg/kg/d	3-4	liquid	$
Naproxen	15 mg/kg/d	2	liquid	$$
Tolectin	30 mg/kg/d	3	older children	$$
Piroxicam	10-20 mg/kg/d	1	older children	$$
Relafen	1000-1500 mg/d	1	older children	$$$

Juvenile Rheumatoid Arthritis

New Drugs

- COX-2 inhibitors in use in older children, children with GI complications
- TNF receptor blockers (Enbrel) in some limited trials

Juvenile Rheumatoid Arthritis

Adjunctive Therapy

- Occupational and Physical Therapy
- Encourage exercise and activity
- Night splints sometimes used to treat or prevent deformities
- Occasional steroid injections into joints
- Surgery - synovectomy, correction of deformity, orthodontia (for micrognathia)